surgical treatment of DFSPs. In the setting of advanced metastatic or inoperable disease, conventional chemotherapy has not been efficacious; however, targeted molecular therapy with imatinib, a selective tyrosine kinase inhibitor, has been successful as adjuvant treatment in tumors with COL1A1-PDGFB positivity.[1–5]

Although DFSPs have a propensity to recur, they portend a good prognosis in general; distal metastases are uncommon, occurring in 1% to 4% of cases, and deaths are even more rare, occurring in less than 1% of all cases.[1–5] However, 3% to 20% of DFSP have areas of fibrosarcomatous changes, classifying them as DFSP-FS.[1–5] DFSP-FSs are considered "high-risk" variants and have higher rates of local recurrence, distal metastases, and death.[1–5] Metastases, when they do occur, most often involve the lungs, bone, or local lymph nodes and result in death within 2 years. To the authors' knowledge, this is the first ever report of intra-abdominal metastases of this extent originating from a cutaneous DFSP, as well as the first ever reported case of distant metastasis years after surgical resection with negative histological margins.

Conclusion

Dermatofibrosarcoma protuberan is a rare, slow-growing, and poorly understood mesenchymal soft tissue tumor of the skin that constitutes a difficult management problem. Though local recurrence is common, distant metastases and deaths are rare. However, the fibrosarcomatous transformation represents a high-grade variant with a propensity to recur and metastasize. The identification of key clinicopathologic features may influence the choice of treatment and outcome for these patients.

References

1. Criscione VD, Weinstock MA. Descriptive epidemiology of dermatofibrosarcoma protuberans in the United States, 1973 to 2002. J Am Acad Dermatol 2007;56:968–73.
2. Hoesly PM, Lowe GC, Lohse CM, Brewer JD, et al. Prognostic impact of fibrosarcomatous transformation in dermatofibrosarcoma protuberans: a cohort study. J Am Acad Dermatol 2015;72:419–25.
3. Minter RM, Reith JD, Hochwald SN. Metastatic potential of dermatofibrosarcoma protuberans with fibrosarcomatous change. J Surg Oncol 2003;82:201–8.
4. Snow SN, Gordon EM, Larson PO, Bagheri MM, et al. Dermatofibrosarcoma protuberans: a report on 29 patients treated by Mohs micrographic surgery with long-term follow-up and review of the literature. Cancer 2004;101:28–38.
5. Paradisi A, Abeni D, Rusciani A, Cigna E, et al. Dermatofibrosarcoma protuberans: wide local excision vs. Mohs micrographic surgery. Cancer Treat Rev 2008;34:728–36.

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Small Intestinal Submucosal Matrix as a Novel Reconstructive Option for Large Scrotal Defects

The scrotum presents a challenging area to reconstruct while maintaining aesthetic and functional outcomes. Reconstructive approaches that have been used for scrotal defects include split-thickness skin grafts (STSGs), myocutaneous, and fasciocutaneous flaps. Despite these varied techniques, there remains a need for an alternative reconstructive approach that involves a less invasive measure for closure with a better ability for tumor surveillance. Thus, the authors present the use of the porcine small intestinal

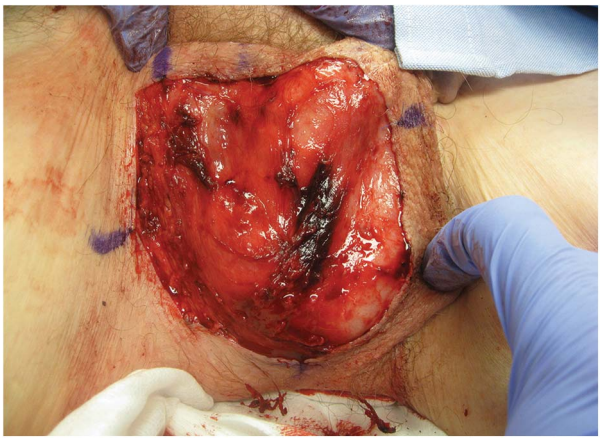

Figure 1. Right scrotal defect extending down to the dartos fascia and covering the entire right scrotum.

Figure 3. Reduction in the right scrotal wound size 4 weeks after small intestinal submucosal matrix placement.

submucosal (SIS) extracellular matrix (Oasis Ultra) as a novel alternative therapy for the repair of large scrotal defects.

An 82-year-old man presented with a 4-year history of an itchy, red, and scaling rash on the right hemiscrotum. Biopsy revealed extramammary Paget disease (EMPD) with an initial lesion size of 3×3 cm. The lesion was resected with negative margins after 4 stages of Mohs micrographic surgery (MMS) leaving a defect of 8.5×6 cm (51 cm^2), covering the entire right scrotum (Figure 1). After resection, the depth of the defect extended to the dartos fascia, which lies immediately deep to the scrotal epidermis and dermis but superficial to the external spermatic fascia. Given the patient's age as well as the location and size of the defect, the wound was left to heal by secondary intention facilitated and accelerated by the placement of a tri-layered porcine SIS extracellular matrix (Oasis Ultra). The matrix was cut and sized to cover the wound surface and to extend slightly beyond the margins, secured with the 5-0 vicryl rapide sutures and rehydrated with sterile saline (Figure 2). The matrix was covered with a nonadherent dressing and left undisturbed for 7 days, after which daily wound care using diluted acetic acid and petroleum ointment was initiated.

At 1-week follow-up, the entire wound base was granulated and by 4 weeks, the wound size had decreased by over 50% to 3.9×3.2 cm (Figure 3). During the first few weeks postoperatively, the patient reported only occasional scant bleeding from the area

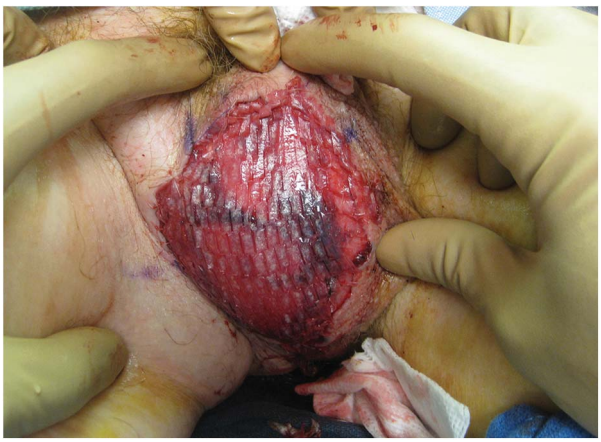

Figure 2. Rehydrated tri-layered porcine small intestinal submucosal extracellular matrix secured to the wound bed.

Figure 4. Excellent functional and aesthetic outcome at 5 months after small intestinal submucosal matrix placement.

during increased physical activity. The wound closed by 8 weeks, and the residual scar was soft and supple at 14 weeks. A long-term follow-up at 5 months revealed excellent cosmetic results (Figure 4). The patient was satisfied with the overall functional and aesthetic outcome. Moreover, patient-reported pain was minimal throughout healing.

The SIS matrix is a native minimally processed natural biomaterial derived from the porcine small intestinal submucosa used to facilitate healing in wounds that have an unfavorable healing environment. The SIS matrix is currently indicated for diabetic ulcers, pressure ulcers, chronic vascular ulcers, and surgical wounds.[1] It provides a 3-dimensional scaffold to aid in the regeneration of tissue where the existing extracellular matrix may be suboptimal for wound healing. Components found in the human dermis including collagen, elastin, glycosaminoglycans, glycoproteins, and proteoglycans are retained in the matrix, making it a dermal substitute that closely resembles the native dermis. The important bioactive factors such as basic fibroblast growth factor and transforming growth factor β are also retained in their active forms in the SIS matrix, which are crucial for stimulating angiogenesis, capillary ingrowth, and tissue regeneration. Another bioengineered product also derived from the porcine tissue is E-Z Derm. Unlike E-Z Derm which is a biosynthetic dressing, SIS matrix is a native, minimally processed extracellular matrix classified as a cellular and tissue-based product that becomes incorporated into the wound. This lies in contrast to E-Z Derm, which serves as a protective dressing but does not incorporate into the wound.

Currently, there are several reconstructive approaches that have been used for scrotal defects. Split-thickness skin grafts are frequently used as they can cover large defects, but are limited in their pliability to adjust to the contour of the scrotum which can lead to a decreased graft take. Myocutaneous[2] and fasciocutaneous[3] flaps have also been reported in the literature as options for scrotal reconstruction, particularly with deeper wounds. Flaps, such as the anterolateral thigh flap, offer the advantage of a large cutaneous area and long pedicle, making it ideal for larger and deeper defects. Chen and colleagues[4] documented a series of 30 cases of EMPD that were treated by STSGs, adjacent tissue flaps (scrotal or low abdominal rotary flaps), or anterolateral thigh flaps. Of the 15 patients treated with STSGs, 4 patients experienced some degree of graft necrosis. There were no serious complications in the 5 patients treated with anterolateral thigh flaps and only 1 reported paroxysmal neuralgia and numbness of the lower limb postoperatively. When choosing which reconstructive modality to use, one must consider the unique anatomical layers of the scrotum. From superficial to deep, these layers include the epidermis, dermis, dartos fascia, external spermatic fascia, cremasteric muscle, internal spermatic fascia, and tunica vaginalis.

In the case of the patient's scrotal reconstruction, the SIS matrix offers the benefit of avoiding invasive reconstruction and increasing the rate of healing through granulation. In EMPD, it allows for improved tumor surveillance, which is significant with this disease, as tumor recurrence rates are 16% for primary EMPD and 50% for recurrent EMPD after MMS.[5] Flaps and STSGs impede tumor surveillance by covering up the native tissue that may still contain tumor cells, which can allow further progression of disease before recurrence is detected. The SIS matrix evades this problem, as it is acellular and allows the ingrowth of native cells into its structure.

In conclusion, the porcine SIS matrix offers an effective alternative reconstructive option for large scrotal defects resulting from the tumor extirpation. Although the use of bioengineered tissue products and dressings in MMS has been reported in other anatomic locations, to the best of the authors' knowledge, this report is the first to describe a Mohs micrographic surgical reconstruction of a scrotal defect using a bioengineered tissue product or dressing. The SIS matrix is ideal for patients who do not desire further invasive procedures and affords better surveillance for tumor recurrence. It has the utility for healing Mohs micrographic surgical wounds postoperatively with excellent functional and aesthetic outcomes in anatomic areas with difficult wound-healing environments and therefore should be considered as a viable option for scrotal defects or other less ideal surgical wounds.

References

1. Beers PJ, Adgerson CN, Millan SB. Porcine tri-layer wound matrix for the treatment of stage IV pressure ulcers. JAAD Case Rep 2016;2: 122–4.
2. Hsu H, Lin CM, Sun TB, Cheng LF, et al. Unilateral gracilis myofasciocutaneous advancement flap for single stage reconstruction of scrotal and perineal defects. J Plast Reconstr Aesthet Surg 2007;60: 1055–9.
3. Aydin T, Feyzi K, Tayfun T, Berna T. Reconstruction of wide scrotal defect using groin fasciocutaneous island flap combined with a strip of deep fascia. J Plast Reconstr Aesthet Surg 2010;63:1394–5.
4. Chen Q, Chen YB, Wang Z, Cai ZK, et al. Penoscrotal extramammary Paget's disease: surgical techniques and follow-up experiences with thirty patients. Asian J Androl 2013;15:508–12.
5. Hendi A, Brodland DG, Zitelli JA. Extramammary Paget's disease: surgical treatment with Mohs micrographic surgery. J Am Acad Dermatol 2004;51:767–73.

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